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Medical Image

Title: Von Recklinghausen's Disease and Gastrointestinal Stromal Tumors

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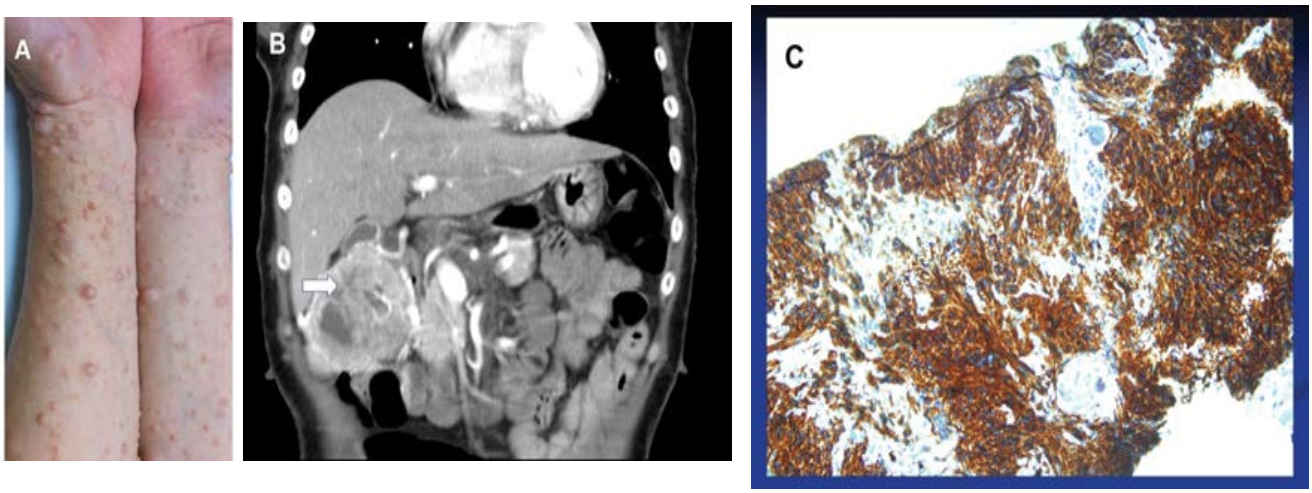

A 51-year-old woman with type 1 neurofibromatosis, also known as von Recklinghausen's disease, presented to the emergent room with melena for several days. The vital signs were normal. Physical examination revealed typical café-au-lait spots all over the body as well as multiple cutaneous and subcutaneous skin nodules (Panel A). Laboratory tests including the tumor markers were normal; however, hematocrit was reported to be 30.5%. An endoscopic examination revealed a gastric ulcer with hemorrhage, which was treated by endoscopic hemostasis treatment. The ultrasonic examination showed a huge intra-abdominal tumor. A computed tomography scan of the abdomen detected a retroperitoneal heterogeneous tumor of 10 cm in size with necrosis inside the tumor (Panel B, arrow). A biopsy procedure was performed and the specimens were sent for pathological examination. H&E staining showed spindle shaped neoplastic cells with whorl like growth pattern. The tumor cells were positive for C-Kit (CD117 leukocyte antigen) immunostain (Panel C). The patient was diagnosed to have a retroperitoneal gastrointestinal stromal tumor. After down-staging with imatinib mesylate, surgical resection was performed five months later. Intraoperative findings identified two 3 cm tumors in the jejunum and a huge 10 cm mass in the retroperitoneal area. The patient has remained in good health without recurrence or metastasis after the surgical procedure.

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